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A peculiar change in the spleen (fibrosis circumscripta lienis).

by Georgy Lubarsky.

Translated from: Zentralblatt fuer Allgemeine Pathologie und Pathologische Anatomie, 65: 291-294 (1936).

Rotter has reported on a peculiar splenic change in Volume 259 of Virchow's Archives, which he called "fibrosis circumscripta lienis." He found a focus in the interior of the spleen, according to him of inflammatory genesis, with the following properties: The size of a dwarf apple, radially constructed, extraordinarily solid, greyish-white on the sectional plane. I had occasion to observe a similar case.

This splenic change was noted as an incidental condition during the section of a 61-year-old man. I extract the following from the autopsic record: Focal pneumonia in the right lower lobe. In the right occipital lobe of the cerebrum, an old focus of softening simulating the signs of porencephaly. Moderate general arteriosclerosis. Moderate general atrophy of the internal organs. The spleen measures 12 X 8 X 4 cm, the capsule is wrinkled, no other pathologic lesions are visible. The splenic parenchyma contains (directly beneath the capsule) a rather sharply delineated bony tumor, extending to the hilus and possessing a bluish-white sectional surface recalling cartilaginous tissue (Fig. 1). On a splenic roentgenogram the tumor shows a shadow whose intensity corresponds to bone tissue and which distinctly reveals the radial structure of the tumor, resembling a coral structure (Fig. 2).

The following histological picture presented itself after lengthy decalcification: The tumor consists of thick, hyaline connective tissue fibers, poor in cells and calcified in spots; now parallel, now forming intersecting bundles, and now with a circular, vertebra-like orientation. The calcified parts may be said to contain no cellular elements at all, whereas the connective tissue in other places contains cells in various amounts, although it is everywhere conspicuously poor in cells. The connective tissue cells are oddly extended, as if they were compressed by the fibers. There are scattered areas of varying size in the tumor that still contain recognizable splenic tissue. In these parts we generally find fairly numerous blood vessels surrounded by small, round cells in an arrangement recalling splenic follicles; the connective tissue penetrates between these, giving the appearance as if the cells occupy only the gaps in the connective tissue. Other regions also reveal rather copious vessels with thickened walls; with narrow but distinct lumina, not infrequently filled with red blood corpuscles. The tumor is not sharply delineated against the normal splenic tissue; it has no capsule, the connective tissue fibers penetrate the splenic parenchyma radially at numerous points, in other places the splenic tissue enters between the connective tissue fibers. The splenic capsule is distinctly visible also above the tumor, moderately thickened; the

tumor does not reach the capsule, however, and is situated in the splenic parenchyma. It is interesting to note that isolated spots, precisely where the hyaline connective tissue has the least cells, often contain large blood vessels whose lumen is filled with erythrocytes and whose walls are completely transformed to hyalin. The various layers of the vascular wall are not recognizable, not even the membrana elastica interna, and the vascular wall fuses gradually with the surrounding hyaline connective tissue. These blood vessels frequently seem to form a large sinus, at least of a size not generally attained by blood vessels in the splenic parenchyma. It is noted that the calcigerous portions apparently form a net --- similarly to the bony trabeculae in the vertebrae, although a true bone formation is not visible anywhere --- and that the non-calcigerous hyaline tissue fills the gaps of this net. It must be stressed that the calcigerous parts contain considerably fewer blood vessels than the other parts, although the connective tissue in some spots of the calcigerous parts has such a circular course that completely closed vascular contours, communicating with the connective tissue, seem to be evident. The round cells deposited especially in the tumor's periphery and in the gaps of the connective tissue, correspond to lymphocyte-like cells; positive inflammatory lesions could not be established, however.

Van Gieson's stain colors the marginal portions of the fibers an intense red, the central and, especially, the calcigerous parts, orange or yellowish red, even yellow. The picture is similar in sections stained with Mallory's fuchsin-aniline red-orange: The marginal fibers of the vascular wall generally are well preserved, thickened here and there, coarse; but we also find vessels in which they are completely or predominantly destroyed. In some places only circularly arranged remains of elastic fibers indicate the past location of a vessel. No ferrous pigments were found in the tumor.

In the sections obtained from the remaining parts of the spleen, the follicles are reduced in size, they frequently consist only of a few rows of cells. The pulpa shows a distinct increase in connective tissue and the picture generally corresponds to that of a senile, atrophied spleen. The vascular walls, especially those of the larger blood vessels in the trabeculae and of the vessels traversing the follicles, are moderately thickened, the lumen is narrowed in places. The splenic pulpa contains a fairly large number of cells with a fine ferrous pigment. No signs of a specific inflammatory process could be found.

Aside from phleboliths, the occurrence of calcified tumors in the spleen is rare. Thus, for instance, tumors (Lubarsch, Fink, Milligk, Kaufmann), infarcts (Lubarsch), tubercles and gummata may scarify and calcify. According to Lancereaux, a calcigeration in the splenic parenchyma is extremely rare and usually is a sequel of infarcts owing to the deposit of a calcified zone around the necrotic tissue. Kaufmann-Wolf report a case of a large calcigerous node in the spleen which was correlated to the scarification of a splenic rupture.

Sternberg describes a case in which a hemangioma calcified in the spleen.

Our case could not be included in one of the above categories except with certain reservations, since the characteristic picture was displaced due to advanced regressive changes (hyaline degeneration, calcigerous precipitation). The splenic change observed by me is closest to Kotter's case, although an important difference in his case is the presence of numerous clods of hemosiderin in the tumor, as well as the demonstration of wide, blood-containing sinuses in the margins of the splenic parenchyma, penetrated by connective tissue strands. In this case calcigerous precipitation was absent. This circumstance caused Kotter to assume an inflammatory genesis of the tumor. These changes were absent in our case and the microscopic picture offered no evidence of an inflammatory origin. The involvement of a calcified gumma, a tubercle or an infarct can be excluded with relative certainty. Tissual changes characteristic of the first two could not be demonstrated; the circumstance that the change does not involve the capsule and that the capsule was entirely unimpaired, even above the tumor, speaks against the possibility of an infarct.

Finally, the genesis of the tumor must be considered. Fibromas and hemangiomas may occur in the spleen and may scarify or calcify. The assumption of fibroma is supported by the fact that the tumor is quite well circumscribed, that it definitely impresses as a tissual increase and that the histologic structure, the bundle-like, crosswise and vertebra-like arrangement of fibers corresponds to a fibrous tumor. The calcigerous deposits naturally are a secondary phenomenon. Whether it we are really dealing with a true tumor, namely a fibroma, or only with a tumor-like proliferation of the connective tissue (a so-called circumscribed fibrosis) is difficult to decide and a decision is quite impossible in our case. The change could be called "fibrosis circumscripta lienis," although this designation gives no clues to the nature of the process, especially its genesis. ( )

Illustrations.

Fig. 1. Cross section through the splenic tumor.
Fig. 2. Roentgenogram of the spleen. The radial structure of the tumor is quite distinct.